Passive ocular proptosis

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SUMMARY Two patients with oculomotor neuropathy demonstrated passive ocular proptosis. In both instances, there was no evidence of a pathological process exerting a vector of force through the orbital opening. The proptosis is caused by a combination of the loss of the normal backward force exerted by the ocular recti muscles and the presence of a small anteriorly directed force exerted by the ocular obliques. Computerised tomography of the orbital and retro-orbital regions was of value in establishing the diagnosis and excluding orbital disease in one patient. The role of computerised tomography in the evaluation of the patient with ocular proptosis is discussed.

Unilateral orbital proptosis (exophthalmos) is usually associated with pathological conditions that exert a vector of force directed externally through the orbital opening. Most commonly, unilateral exophthalmos results from a mass lesion at the orbital rim, such as a lacrimal gland tumour, a mass behind the globe but within the orbit, such as an optic nerve glioma, or a retro-orbital mass, such as a sphenoid wing meningioma. However, the proptosis may rarely be seen as a passive phenomenon secondary to impairment of the supporting tissues of the globe. The following patients are examples of passive ocular proptosis due to oculomotor nerve palsy and illustrate the use of computerised tomography (CT scan) in the evaluation of exophthalmos.

Case reports

CASE 1

A 55 year old man, chronically ill from severe diabetes mellitus, chronic renal failure, and coronary atherosclerotic heart disease, experienced sudden, throbbing, right sided supraorbital and retro-orbital head pain. This was followed within 24 hours by total closure of the right eye and, when the eyelid was passively raised, diplopia. Although requiring, and taking, insulin, his diabetes was in poor control just before the onset of symptoms. He was otherwise well without

recent history of fever, nausea, vomiting, photophobia, trauma, or facial infections. He was admitted to the Ann Arbor Veterans Administration Hospital three days after the onset of his symptoms.

Approximately 18 years earlier, the patient had a similar episode, marked by headache, ptosis, and diplopia which resolved after several weeks. His diabetes was first diagnosed at that time and has since required insulin maintenance. The patient had no other eye complaints except gradually decreasing visual acuity, corrected by refraction, and fluctuating worsening of his visual acuity when hyperglycaemia was pronounced.

On neuro-ophthalmological examination, the patient demonstrated 4 mm of proptosis, directed anterolaterally (Fig. 1) without tenderness to movement or palpation, and there was normal retropulsion. No bruits, pulsations, or venous engorgement were noted. Complete ptosis was present at rest but 2 mm of levator function was performed. Central vision was without scotoma or visual field defect. Cataracts and a non-proliferative retinopathy were present bilaterally. No optic nerve changes were noted. Corneal and periorbital sensation was normal. Tears and periorbital sweating were symmetrically present. Pupillary reactions, both direct and consensual, were present equally well. Spontaneous ductions were full in the left eye. The only spontaneous ductions present in the right eye were those performed by the lateral rectus and the superior oblique muscles. All other motions of the right eye were absent. Forced ductions of the right eye

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Accepted 16 July 1977

Fig. 1 Case 1. Superior photograph showing anterior-lateral displacement of right eye.

were normal. An edrophonium test (10 mg edrophonium chloride, intravenously) gave no change in the eye findings in the presence of systemic pharmacological responsiveness.

Radiographs of the skull and sinuses were normal. A lumbar puncture was performed to exclude subarachnoid bleeding and an acellular cerebrospinal fluid sample was obtained. Blood sugar at admission was 18.26 mmol/l (329 mg/dl). Two consecutive sedimentation rates were elevated (55 and 56 mm/h, Wintrobe) and a biopsy sample of the main temporal artery was normal. The clinical impression was diabetic cranial neuropathy of the right oculomotor nerve.

CASE 2

A 75 year old man suddenly developed ptosis and diplopia at age 72 years. The diplopia, however, was noted only when the ptosed lid was lifted; the patient's vision was otherwise essentially monocular. A vague headache accompanied the onset, but there was no eye pain or constitutional symptoms. Similarly, there were no identifiable precipitants or paralleling systemic disease processes. Over the subsequent three years there has been gradual anteromedial displacement of the globe and persistent diplopia. The ptosis has partly improved thus allowing binocular vision. He was referred to the Ann Arbor VA Hospital at age 74 years after symptoms of weakness, weight loss, and a change in the colour and texture of skin

and hair appeared. Skull radiographs and bilateral carotid angiography were performed before referral and were normal. Further radiographs of the skull, orbits, superior orbital fissures, and tomography of the sella turcica were normal, as was a brain scan. Panhypopituitarism was suggested after endocrinological evaluation demonstrated subnormal values for thyroid, adrenal, and gonadal function tests. The probability of panhypopituitarism was affirmed by abnormal provocative tests and near normal values for hypothalamic releasing hormones. Metyrapone administration failed to elevate diagnostically above baseline the plasma cortisol values or the urinary excretion of 17-ketosteroids and compound S. Fasting blood sugar and a three hour glucose tolerance test were normal. Appropriate replacement therapy was begun.

He was readmitted to the Ann Arbor VA Hospital one year later at age 75 years where stability of his endocrinological status on replacement therapy was documented. The preceding radiological studies were repeated and were again normal. Computerised axial tomography of the head was performed with particular emphasis on the orbital and retro-orbital areas (Fig. 2). No periorbital, intraorbital, or retro-orbital abnormalities were noted either before or after infusion of contrast medium. Several 4 mm sections were made at the level of the orbits to define carefully the appearance of the optic nerves and extra-

Fig. 2 Case 2. CT scan (non-enhanced) of the orbits at level of optic nerves. Periorbital, orbital, and retro-orbital areas are normal. A very slight asymmetry of globe positions is noted. The dissimilar lens images suggest dysconjugance.

Fig. 3 Case 2. Superior photograph showing slight anterior displacement of left eye.

ocular muscles. Only a small degree of anterior displacement of the left globe, and dissimilar lens images, suggesting dysconjugance, were seen.

The neuro-ophthalmological examination showed the same abnormalities on his two Ann Arbor VA hospitalisations, and are summarised as follows. The general appearance of the patient was marked by 2 mm of proptosis (Hertel exophthalmometer) of the left eye with normal retropulsion (Fig. 3). There was no pulsation, bruit, or venous engorgement present. Four millimetres of ptosis were present at rest, as well as 4 mm of levator function. Visual acuity (with refraction), one metre tangent screen examination, and funduscopy evaluation were normal. The corneal and periorbital sensation was normal. A dilated, fixed pupil was present on the left. Normal pupillary responses and spontaneous ductions were present on the right. Spontaneous ductions of the left eye showed the functions of the lateral rectus and superior oblique to be normal, whereas the other recti and the inferior oblique were moderately restricted in movement. Forced ductions of the left eye were normal, and an edrophonium test (10 mg edrophonium chloride, intravenously) gave no change in the eye findings. The clinical diagnosis was pituitary apoplexy with oculomotor nerve infarction.

Comment

Both these patients clinically had oculomotor nerve infarction as evidenced by the suddenness of onset, totality of the ptosis, the paresis of oculomotor nerve innervated extraocular musculature, and the normality of both fourth and sixth nerve function. The sparing of pupillary function and the peri- and retro-orbital pain in the first patient, a severe diabetic, is typical of diabetic oculomotor neuropathy (Dreyfus *et al.*, 1957). Angiography to exclude the rare posterior communicating artery aneurysm that spares the pupil (Kasoff and Kelly, 1975) was not thought to be indicated clinically without evidence of subarachnoid haemorrhage and because of the patient's general state of health. Temporal arteritis, although a consideration and probably mediated by a microvascular infarction of the oculomotor nerve (Walsh and Hoyt, 1969b) similar to that of diabetes, was probably excluded by the biopsy of the trunk of the main temporal artery (Meadows, 1966). Therefore, high dose corticosteroids in spite of the negative biopsy was judged particularly hazardous because of his precarious diabetic control, even though some investigators recommend therapy in the face of a

negative temporal artery biopsy (Hunder *et al.*, 1969).

The second patient's total ophthalmoplegia and panhypopituitarism were temporally related and probably represent the form of adenoma described by Symonds where a tongue of the adenoma escapes from the sella turcica and compresses the third nerve against the dura mater near its entrance into the cavernous sinus (Symonds, 1962). According to Symonds, this tongue of the adenoma may become strangulated and produce infarction, thus resulting in a sudden third nerve palsy or total ophthalmoplegia and hypopituitarism. Both patients, therefore, represent forms of oculomotor neuropathy, one manifestation of which was passive proptosis.

The four rectus muscles normally exert a retraction effect on the globe. Their long courses from the posterior pole of the orbit mediate a force directed backward that is normally countered by the anterior displacement effect of the oblique muscles. In oculomotor nerve lesions, three of the four recti may be involved to produce up to 3 to 4 mm of proptosis (Walsh, 1957). This effect is presumably a combination of the loss of the normal retraction effect of the three recti and the active anterior and inferior displacing effect of the superior oblique. Similarly, in the neuromuscular paresis of myasthenia gravis, ocular proptosis may occur (Walsh and Hoyt, 1969a) which has been observed to disappear after neostigmine administration (Hatch, 1952). In this latter instance, the degree of proptosis and the axis of deviation will depend on the profile of individual musculature impaired by the defective neuromuscular transmission.

The separation of disorders that produce proptosis secondarily, from those groups of diseases that produce proptosis actively (anteriorly directed vector of force) is usually not difficult. By noting its frequent occurrence with oculomotor neuropathy, a puzzling finding may be explained and, when there is no other contrary evidence, may spare the patient unnecessary diagnostic tests or diagnostic exploration. However, the occurrence of proptosis in a diabetic, for example, still necessitates a search for an active process such as cavernous sinus thrombosis. The separation of processes is made easier by the presence or absence of certain clinical signs. In these two patients, normal retropulsion and the absence of eye tenderness, chemosis, dilated episcleral veins, pulsations, and bruits all indicated the absence of a force generating lesion. The forced duction test (FDT) determined that the disturbance in ocular motility was not due to a mechanical

obstacle (Von Noorden and Maumenee, 1967). In thyroid eye disease, for example, the FDT can establish that the limitation of upgaze is due to a restricted inferior rectus and thus correlate with the proptosis, and other physical signs of thyroid eye disease. Similarly, limitation of the FDT will be seen in processes involving the muscle cone near the superior orbital fissure, whereas oculomotor neuropathies and myasthenia gravis are associated with a normal FDT since there is no impedence to muscle motion.

Computerised axial tomography (CT scanning) is now felt to be the diagnostic procedure of choice in evaluating exophthalmos. Particularly with the use of contrast enhancing infusions, and the capability of thin tomographic sections, an accurate definition of the orbits and its contents can be obtained (Lampert *et al.*, 1974). Separation of the images produced by the inferior rectus muscle and the optic nerve can now be made more clearly with these refinements in CT scanning, and the difficulty in differentiating optic nerve tumours from thyroid eye disease is becoming less common. Periorbital tumours such as a lacrimal gland tumour, and retro-orbital tumours, such as sphenoid wing meningiomas, are also easily detected by CT scanning. In the second patient, the normal CT scan, along with other data, corroborates the clinical impression of passive proptosis after oculomotor nerve infarction.

In summary, not all protrusions of the globe are due to expansile disease within or near the orbit. Proptosis may occur passively and may indicate a disease process far different from those usually considered in the differential diagnosis of exophthalmos. Myasthenia gravis and oculomotor nerve lesions are the most likely causes of this passive proptosis.

The cooperation of Dr Joachim F. Seeger of the Department of Radiology, University of Michigan Medical Center, is gratefully noted. I am apprecia-

tive of the photographic assistance of Robert J. McKnight, Chief, Medical Media Production Service, Ann Arbor Veterans Administration Hospital. Dr Russell N. DeJong, Chairman Emeritus, Department of Neurology, University of Michigan Medical Center, kindly read the manuscript and offered constructive comments.

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